SPINE Volume 19, Number 4, pp 463–465

Monostotic Fibrous Dysplasia of the Thoracic Spine

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Fibrous dysplasia involving the thoracic spine is very rare. This patient had monostotic fibrous dysplasia involving the vertebral body, posterior elements, and right seventh rib with focal neurologic signs. The radiographic and bone scan findings are shown. The patient was treated successfully with tumor resection and arthrodesis using combined anterior and posterior approaches. Although fibrous dysplasia is classified as a benign process, clear margins of resection are required to prevent recurrence. [Key words: fibrous dysplasia, thoracic spine, resection-arthrodesis]

Fibrous dysplasia involving the thoracic spine, in either its monostotic or polyostotic form, is very uncommon. The majority are discovered as incidental x-ray findings, and the lesions are usually asymptomatic.

A case of symptomatic thoracic spine involvement with focal neurological signs that was treated successfully with resection and arthrodesis is presented.

Case Report

A 46-year-old woman, working as a process worker, who was otherwise well presented to her local doctor with palpitations.

A chest x-ray, done as part of the investigations, showed an expanding lesion in the posterior aspect of the right seventh rib and adjacent vertebral body (Figure 1). The only symptom referable to her back was mild intermittent pain in the thoracic region, aggravated by activity and relieved by rest. Nothing significant was found in the patient's family or past medical history.

Clinical examination revealed a low, diffuse paravertebral swelling at the right seventh thoracic vertebral level with no neurologic deficit. Chest auscultation was normal, and there were no soft-tissue lesions or cafe-au-lait spots. Complete blood count, sedimentation rate, serum calcium, and alkaline phosphatase were all normal.

Bone scan (Figure 2) showed markedly increased uptake in the right seventh rib and adjacent vertebra consistent with increased bone activity. On computerized axial tomography (Figure 3), the posterior aspect of the seventh rib was grossly expanded, with the lesion crossing the costochondral junction, involving both the body and posterior elements of the seventh thoracic vertebra, eroding through into the spinal canal. Myelography confirmed no spinal cord or nerve root compression.

Our differential diagnosis included fibrous dysplasia, osteoblastoma, eosinophilic granuloma, aneurysmal bone cyst, giant cell tumor, and chordoma or solitary metastasis. A trephine biopsy of the seventh thoracic vertebra confirmed the diagnosis of fibrous dysplasia.

During the next 6 weeks, the patient developed increasing pain in the thoracolumbar spine and right chest posteriorly, with mild sensory changes in the right seventh thoracic nerve root, and surgical treatment was advised.

Initially, a T5 to T9 posterior spinal fusion, using TSRH instrumentation (Danek Group Inc., Memphis, TN) with transverse process hooks and cross linkages to ensure stable fixation and autogenous iliac crest bone graft, was performed. The posterior elements of the seventh thoracic vertebra were porous, distorted, and very soft, and a right laminectomy, costotransversectomy, and pediculectomy were performed. A right thoracotomy was performed, resecting both the sixth and seventh ribs. A partial seventh thoracic vertebrectomy was also performed. A rib graft was placed anteriorly in the defect, which was also packed with autogenous iliac crest bone.

The patient made a full and uneventful recovery, mobilized at 2 weeks in a Taylor brace, and was discharged 4 weeks after the operation, walking independently with one walking stick.

At 6 months after the operation, she was asymptomatic and able to do light housework, with x-rays showing solid anterior fusion (Figure 4).

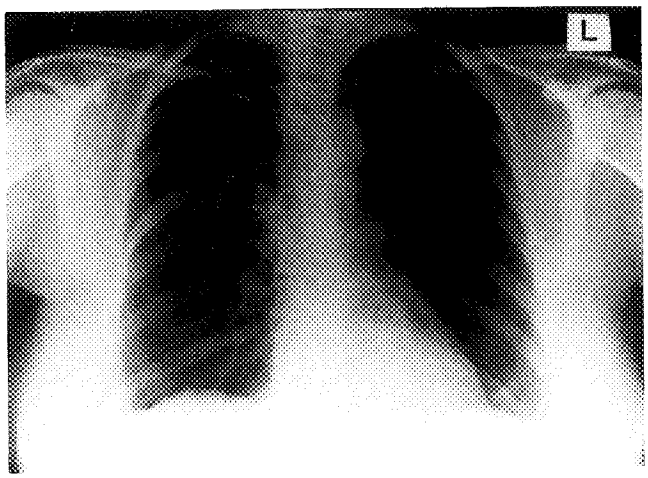

Figure 1. Chest radiograph showing expansile lesion involving right seventh rib and seventh thoracic vertebra.

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Accepted for publication March 5, 1993.

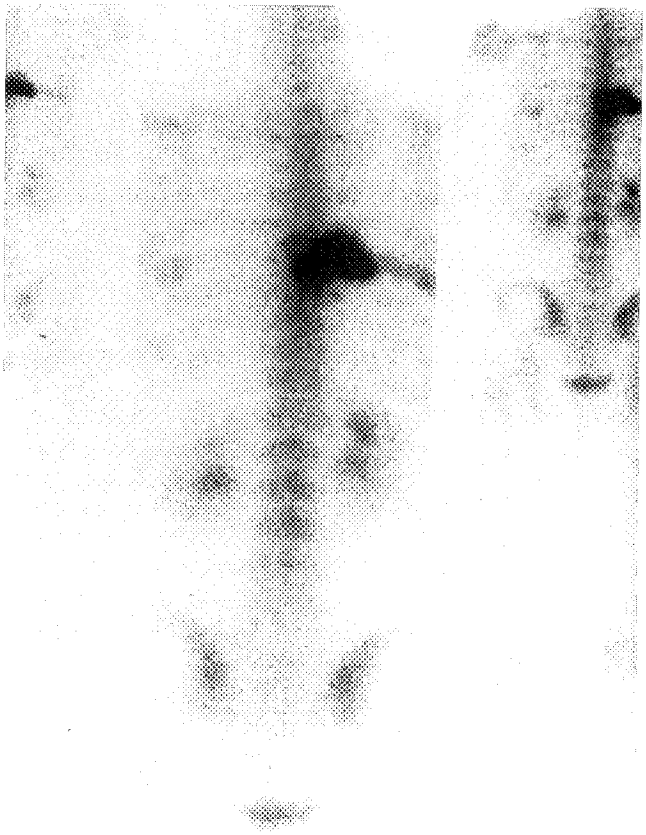

Figure 2. Bone scan demonstrating markedly increased uptake of seventh thoracic vertebra.

Discussion

Fibrous dysplasia is a benign, relatively common fibro-osseous lesion of bone, accounting for 2.5% of all bone lesions and 7% of benign bone tumors.[6] The term was first coined by Lichtenstein and Jaffe in 1942,[5] and is believed to represent a developmental abnormality of bone forming mesenchyme. The process may be monostotic, polyostotic, or polyostotic, with cutaneous, endocrine, or both abnormalities (McCune-Albright's Syndrome).

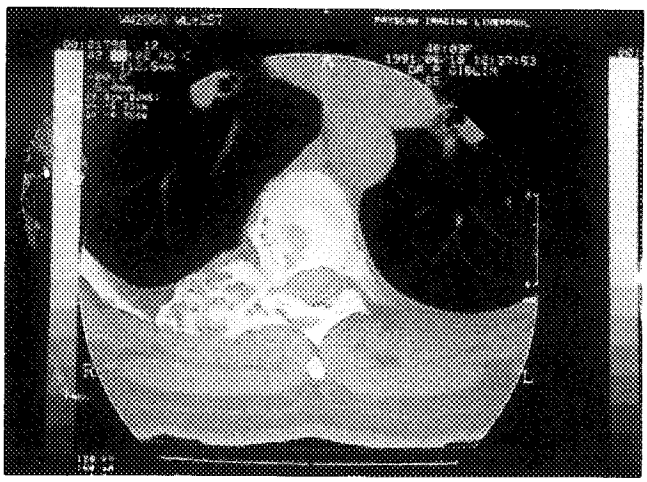

Figure 3. A CT scan of the involved vertebra and rib illustrating the destructive nature of the lesion.

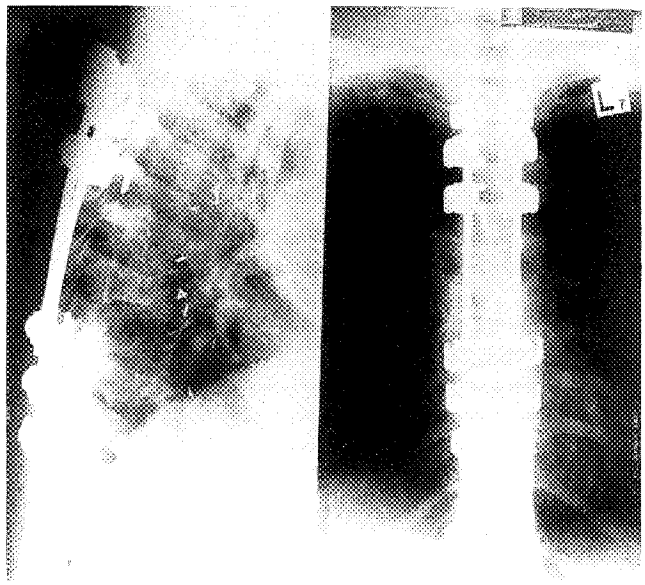

Figure 4. Plain AP and lateral x-rays of thoracic spine at 6 months after operation showing incorporation of the iliac crest and rib grafts.

The monostotic form occurs more frequently than the polyostotic form, with an equal sex incidence. It commonly presents in the first and second decades of life, and neither regresses nor enlarges after the cessation of bone growth.[6] There are no extraskeletal or endocrine abnormalities, and laboratory investigations are invariably normal. Patients are usually asymptomatic, and the lesion is often discovered incidentally. The most common presenting symptom is swelling or deformity, especially in the face, ribs, or lower limbs.

The monostotic form occurs more frequently in the metaphyses of long bones and ribs, and vertebral involvement is rare, with only nine cases reported in the literature. Only two cases affecting the thoracic spine have been described.[6,7]

Rosenblum et al[6] reported a case affecting the body, right pedicle, superior articular facet, and lamina of the first thoracic vertebra and the right inferior articular facet of the seventh cervical vertebra. The patient underwent resection of the mass posteriorly, and then, as a second procedure through a paratracheal approach, excision of the mass from the first thoracic vertebral body and anterior fusion with an iliac crest strut.

Singer et al[7] reported a case involving the 11th thoracic vertebral body, adjacent to the right 11th rib, which had been resected 19 years previously for fibrous dysplasia. This patient underwent posterior stabilization with Cotrel-Dubousset instrumentation, and then, one week later, near-total vertebrectomy and anterior fusion with rib graft.

Henry[2] showed that, to achieve successful surgical treatment, all of the dysplastic tissue must be excised. If any remaining abnormal tissue is in an active state, it will continue to grow and lead to recurrence. He also stated that reactivation may occasionally occur later in life, and this may account for the vertebral involvement in Singer et al's patient many years after the initial resection.

Fibrous dysplasia is a benign bone lesion without destructive capability, and, traditionally, once the diagnosis of monostotic fibrous dysplasia of the spine is confirmed on biopsy, no further treatment is required.[6] Stephenson[8] had four patients in his series with lesions of the spine and pelvis, all of whom were managed conservatively. In more recent reports, resection and arthrodesis have been carried out for spinal lesions with persistent pain, focal neurologic findings, or both.[4,6]

Stirrat et al[9] reported that rare sequelae of spinal involvement are angular deformity, vertebral collapse, and cord compression secondary to posterior expansion of the fibrous tissue mass—these are absolute indications for surgical intervention. In our patient, the decision to surgically excise the lesion was based on the development of progressively increasing thoracolumbar pain and right-sided seventh thoracic nerve root symptoms.

Combined anterior and posterior approaches with fusion are often used in the management of spinal tumors, especially metastases, and, as in this case, they enable the removal of all dysplastic tissue with maintenance of spinal stability.

In all the previously reported surgically treated cases, the posterior and anterior fusions were done as two-stage procedures, usually with 1 week between operations. In this case, both anterior and posterior fusions were performed under one anesthetic without any adverse effects. Although the risk of malignant degeneration is very small, complete excision of the dysplastic tissue was performed to minimize this risk. Histology of the rib and vertebral specimens revealed no evidence of malignancy.

Conclusion

Monostotic fibrous dysplasia involving the thoracic spine is very rare, and has only been described in two cases.[6,7]

We describe a case involving the right seventh rib and adjacent thoracic vertebra in a 46-year-old woman who, after an asymptomatic presentation, developed increasing pain and focal neurologic symptoms.

She was successfully managed with combined posterior and anterior spinal fusions under one anesthetic. Although fibrous dysplasia is a benign process, it does cross joints, and the surgeon must ensure clear margins of excision to prevent recurrence.

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